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Treatment and Outcome in patients with Infantile spasms and Lennox Gastaut Syndrome

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Treatment and Outcome in patients with Infantile spasms and Lennox Gastaut Syndrome

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ABSTRACT:

AIM:

To measure and assess the efficacy of ACTH and Vigabatrin in Infantile Spasm and their Unresponsiveness in Lennox-Gastaut Syndrome in Pakistani Population

MATERIALS AND METHODS

A study was conducted between year 2006 to 2016 evaluating 120 patients presenting to Neurodiagnostic Centre, Hamdard University Hospital and National Medical Centre, Karachi. Infantile spasms and Salaam Attacks were the basic diagnosis both clinically and electroence phalographically. Age ranges between 1 month to 1.5 years. All patients were administered appropriate doses of Anti Epileptics, ACTH and vitamin supplements

RESULTS

Amongst 120 patients of Infantile Spasm that we have encountered in last decade about 98 patients (81.6%) who responded well to treatment were below 11 months.3 patients (2.5%) didn't come for the follow up. 20 (16.6%) patients were totally un responsive to treatment, those patients that didn't respond to full therapeutic doses of appropriate Anti Epileptic and ACTH ranged from ages between 12 months to 1.5 years. Even during the treatment, the EEG pattern transformed from classical Hypsarrythmias in some and Burst Suppression in others to classical 1.5 to 2.5 spikes/sec reflecting Lennox Gastaut Syndrome and are best treated under 1-11 months of age.

CONCLUSION

In view of late arrival of patients, awareness needs to be created so that infantile spasms are picked up at the right time. Patient who have been brought in later stages (>1 year) were unresponsive to treatment and were evolved into Lennox Gastaut Syndrome.

INTRODUCTION:

Infantile Spasms and Lennox Gastaut are severe types of epilepsies in children. It occurs in roughly 2-3 per 10,000 live births3. It usually occurs between 3 and 8 months of age with a peak at 4 months. A triad of Hypsarrythmia, Psychomotor retardation and Epileptic spasms (axial contraction in flexion or extension in clusters is named as West syndrome. Lennox Gastaut Syndrome develops between 2 and 8 years of age and consists of different seizures types. The EEG shows 2.5 Hz of slow spike waves, generalized with bifrontal predominance and burst of rapid rhythms during slow sleep 1. Infantile spasms have many causes but the exact pathophysiology is not yet known. The causes are classified as pre natal, natal and post natal. Amongst 40% of the cases prenatal causes includes Central Nervous System malformations, chromosomal aberrations, TORCH infections and less commonly inborn errors of metabolism, natal causes includes Hypoxic ischemic encephalopathy and hypoglycemia2. Cryptogenic Infantile spasms accounts for most of the cases which lacks a known cause. According to US consensus report on infantile spasms that cryptogenic infantile spasms are those that occurs in context with normal development without a clear etiology[3]

MATERIALS AND METHODS

A study was conducted between year 2006 to 2016 evaluating 120 patients presenting to Neurodiagnostic Centre, Hamdard University Hospital and National Medical Centre, Karachi. Patients were clinically diagnosed as Infantile spasms and Electroencephalography showed definitive evidence of hypsarrythmias. Out of 120 patients 80 patients were male and 40 were females. Age ranges between 1 month to 18 months. ACTH and Vigabatrin combination was the most common treatment together with other anti epileptics like valproate and leviteracitam. Patient who were brought in under 1 year of agere also addressed with requisite medications. started showing response to treatment in 1st few months. This treatment combination also helped improved mental and physical milestones. Children who presented with mixed type of seizures responded less promptly then those with myoclonus only. Few patients (20%) who couldn't afford ACTH were subjected to oral steroids and the outcome was much poorer than that of ACTH. To add to the above, Vigabatrin was used in 70% of cases and found to

be more beneficial than ACTH alone. Most patients were supplemented with pyridoxine and folic acids. Sleep issues were also addressed with requisite medications.

RESULTS

Amongst 120 patients of Infantile Spasm that we have encountered in last decade about 98 patients (81.6%) who responded well to treatment were below 11 months.3 patients (2.5%) didn't come for the follow up. 20 (16.6%) patients were totally un responsive to treatment, those patients that didn't respond to full therapeutic doses of appropriate Anti Epileptic and ACTH ranged from ages between 12 months to 1.5 years. Even during the treatment, the EEG pattern transformed from classical Hypsarrythmias in some and Burst Suppression in others to classical 1.5 to 2.5 spikes/sec reflecting Lennox Gastaut Syndrome and are best treated under 1-11 months of age.

CONCLUSION

In view of late arrival of patients, awareness needs to be created so that infantile spasms are picked up at the right time. Patient who have been brought in later stages (>1 year) were unresponsive to treatment and were evolved into Lennox Gastaut Syndrome.

INCLUSION CRITERIA

- Patients who were less then 2 years
- Evidence of hypsarrythmias in Electroencephalographic findings
- Clinical evidence of Salaam Attacks

EXCLUSION CRITERIA

- Patients who were more than 2 years were excluded

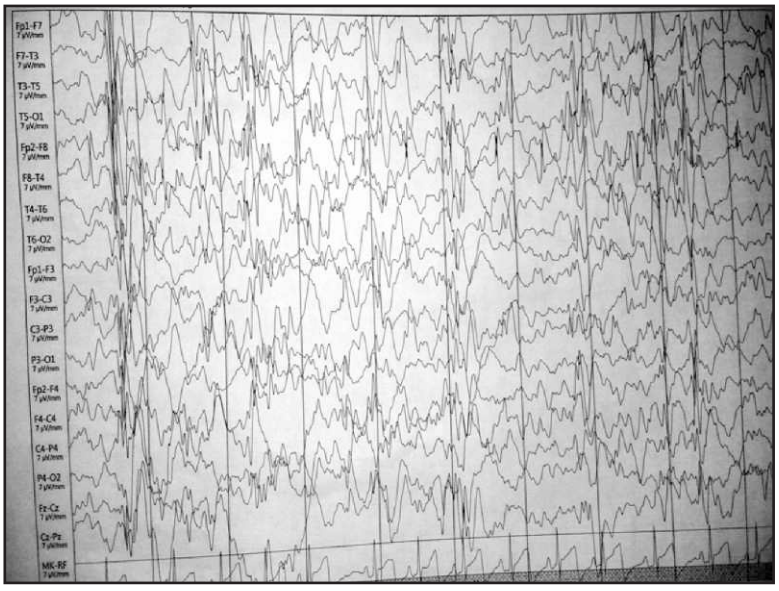

Figure 1
Showing multifocal sharp and spike waves in hypsarrythmia

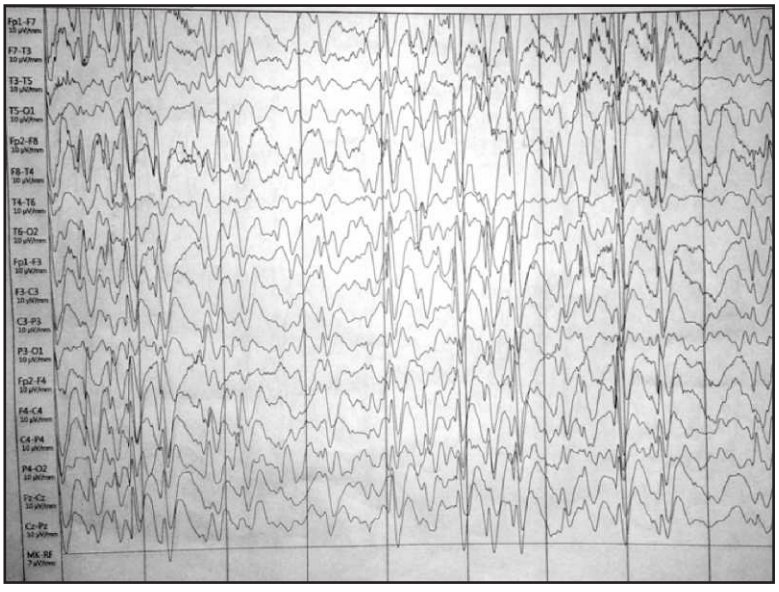

Figure 2
Showing Spike Burst Pattern

DISCUSSION

According to a study done by Chang Yong Tsao on 9 patients who were recently diagnosed with encephalopathy and infantile spasms were given a combination of ACTH and Vigabatrin. ACTH was stopped after 4-6 weeks and infants were given vagabatrin only. There was a complete cessation in all 9 patients[4].

In another placebo-controlled study of vigabatrin in infantile spasms, there was a huge decrease in number of cases that was treated with vigabatrin as compared with the placebo at the end of double blind phase [5].

A randomized study with 14 days ACTH was compared with 14 days of oral steroids. High dose of ACTH was more beneficial in eradicating hypsarrythmias in Electroencephalographic findings and infantile spasm clinically [6].

Ketogenic diet is also beneficial in controlling epilepsy however, the exact mechanism is still not known. A retrospective study was done on 23 patients with age that ranged between 5 months and 24 months were started on a ketogenic diet and as a result, the seizures was improved for more than 90%[7].

Surgical treatment is also beneficial in treating infantile spasms. Positron Emission Tomography was performed on 4 infants with cryptogenic infantile spasms which revealed focal cortical dysplasia which was surgically resected. All infants had resolution of spasms after the surgery[8].

This fact also suggest that the diagnosis and awareness of Infantile Spasm has to be promoted amongst the Pediatricians, the newer Pediatric Neurologists and all those family physicians who are first to encounter these patients

Treated in time with even intensive Salaam Attack can recuperate totally with good treatment given in the nick of time. This will prevent a total loss of mortality amongst infants

Our study includes cases of LG syndrome with myoclonus, abnormal EEG, mental backwardness and behavioral decline

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Arif Harekar; concept, data collection, data analysis, manuscript writing, manuscript review
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